Anesthetic Delivery Management of Multi-organ Systemic Scleroderma

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Effect of SSc on pregnancy

- Affects multiple organ systems
- Increased frequency of miscarriages and preterm deliveries
- Increased maternal and fetal risks
- Risk of hypertensive disorders is controversial
- Challenges in managing airway, hemodynamic, vascular access
- Certain antibodies associated with complications and worse outcomes
- Medication adjustments

Effect of pregnancy on SSc

- Generally well-tolerated
- Stable disease in majority
- Physiological changes exacerbate existing major organ dysfunction
- Cardiovascular and pulmonary considerations
- Improvement of peripheral vascular symptoms observed

Preconception counseling
Frequent monitoring
Multidisciplinary optimization



34-year-old G1P0 with SSc, severe interstitial lung disease (FVC 39%), pulmonary hypertension (RVSP 38 mmHg), Raynaud's, esophageal dysmotility



CT chest - stable chronic diffuse interstitial lung disease consistent with pulmonary manifestations of scleroderma and fibrosis

Pregnancy Course

- Aspiration pneumonia, worsening dyspnea
- 2-4L home oxygen
- Scl-70 high risk of progression
- Deferred immunosuppressive therapy
- Low dose aspirin and prophylactic enoxaparin
- Planned C-section <37 weeks
- C-section at 34 weeks for fetal heart rate decelerations
- Stable intrapartum course
- ICU to floor on POD1
- Neonate: NICU for CPAP and prematurity
- Persistent dyspnea after discharge
 - Invasive mucinous adenocarcinoma

Anesthetic Management

- Skin thickening
 - Thyromental distance < 4 cm
- Poor mouth opening
- Orthopnea, GERD
- Dural puncture epidural
 - Small increments of lidocaine w/ epi, bicarb
- Vasopressin 0.02 units/min
- Stable on 4L NC
- Crystalloids 1L
- UOP 100 mL
- QBL 845 mL

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Management

Pre-pregnancy

- Risk stratification
 - Pregnancy generally not recommended for FVC<50%
- Organ involvement
- Advise on maternal and fetal risks
- Medication adjustment

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- Lack safety data
- Antibody screening

Antepartum

- Increased cardiac and respiratory workload
- Serial TEE, labs, exams
 - PVR rise, RV function
- Home BP
 - Monitor for SRC
- SpO2 >94%
- Monitor GI symptoms
- Medication limitation
 - PAH treatment
- Increased fetal monitoring

Delivery

- Mode of delivery
- Early anesthesia evaluation
 - Airway risks
 - Aspiration precautions
- Pulmonary &CardiovascularConsiderations
- GETA vs Neuraxial
- Vascular access
- Avoid excessive fluid shifts
- Avoid hemodynamic fluctuations
- Anticoagulation

Postpartum

- Strict fluid balance
- Disease exacerbation up to 10%
- Medication resumption
- Fetal outcomes favorable
 - Common complications
 - Autoantibody transfer



^{1.} Roberts, James G., et al. "Progressive Systemic Sclerosis: Clinical Manifestations." J. Clin. Anesth., vol. 14, no. 6, 2002, pp. 474-477. Elsevier, https://doi.org/10.1016/j.jclinane.2022.06.024.

^{2.} Daraz, Yasmeen, et al. "Pregnancy in Pulmonary Arterial Hypertension: A Multidisciplinary Approach." J. Cardiovasc. Dev. Dis., vol. 9, no. 6, 2022, p. 196. MDPI, https://doi.org/10.3390/jcdd9060196.

^{3.} Grant-Orser, Amanda, et al. "Pregnancy Considerations for Patients With Interstitial Lung Disease." CHEST, vol. 162, no. 5, 2022, pp. 1093-1105. https://doi.org/10.1016/j.chest.2022.06.024.

^{4.} Lazzaroni, Maria-Grazia, et al. "Reproductive Issues and Pregnancy Implications in Systemic Sclerosis." Clin. Rev. Allergy Immunol., vol. 64, 2023, pp. 321-342. https://doi.org/10.1007/s12016-021-08910-0.